A Game of Angles: The Importance of the Clival Canal Angle

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Educational Objectives

•Review the clival canal angle

•Convey clinical importance of assessing the clival canal angle, particularly with regards to Chiari I Malformations and implications for corrective surgery

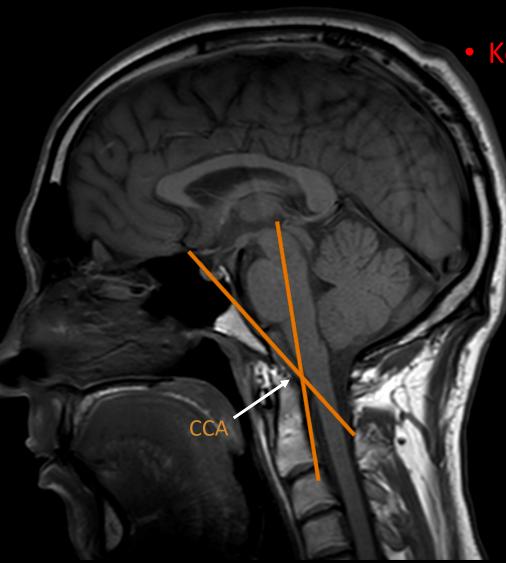
•Highlight flexion MRI as a helpful tool in confirming kyphotic CCA role in bulbar symptoms

 Provide a reference for key points related to clivus canal angle pathology



Clival Canal Angle

- Normal CCA in:
 - Neutral position
 - ~160-170 degrees
 - Flexion- Decreases by up to 10 degrees
 - ~150 degrees
 - Extension- Increases by up to 10 degrees
 - ~180 degrees



Key Point:

- Kyphotic CCA < 150 degrees may be associated with
 - Ventral cord compression
 - Atlanto-dental instability / atlanto-axial subluxation
 - Trauma
 - Developmental and/or heritable hypermobility syndromes
 - Degenerative conditions

Equal to or less than 135 degrees is far more likely to be symptomatic



Abnormal CCA



Abnormal CCA; recurrent dizziness

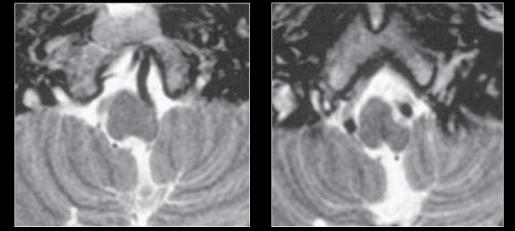
Sagittal MRI demonstrates a kyphotic CCA at 135 degrees. The skull base angle was within the normal range, though at the lower limits of normal (not shown).

Equal to or less than 135 degrees is far more likely to be symptomatic



Cervicomedullary Junction Compression

- Cervicomedullary junction (CMJ) compression is a specific subset of CVJ abnormalities
 - Often associated with a dolichoectatic vertebrobasilar system, CVJ mass, abnormal CCA
 - Vascular compression most commonly occurs along the anterolateral surface of the medulla
 - Corticospinal tracts reside within the lateral segments of the medulla
- Key Point: Even in the absence of a suggestive clinical history, CMJ compression is worth noting as other serious complications include:
 - Posterior circulation TIAs
 - Ischemic or hemorrhagic infarct



a and b. 32 year old female with recurrent throbbing headaches, vertigo, and syncope. T2 images demonstrate a tortuous left vertebral artery impinging upon the left anterolateral surface of the medulla, resulting in posteromedial mass effect.



68 year old male with acute onset ataxia and transient vertigo. Axial T2_{gy} image demonstrating severe compression along the left anterolateral medulla resulting in rightward deviation.

Kyphotic CCA



MRI C-spine obtained for 29-y/o female with a 9-month history of bilateral synchronous, recurrent upper extremity tingling and numbness, always associated with neck and shoulder pain.

Kyphotic CCA at 112 degrees with moderate kinking at the CMJ. No spinal canal or foramina stenosis. No other imagining findings to explain the cause of the patient's symptoms. Imaging findings and symptoms are compatible with CMJ syndrome/referable bulbar symptoms as reported in the literature (Neurosurg Rev (2018) 41:149-163).

Incidentally noted diffusely diminished T1 marrow signal intensity, which in this case was due to red marrow reconversion.



Clinical Importance of Kyphotic CCA

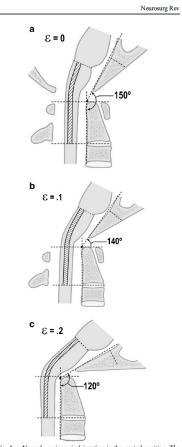
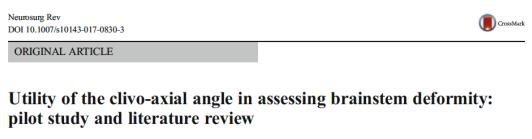


Fig. 1 a Normal craniocervical junction in the neutral position. The CXA varies from 150° to 165°. There is minimal or zero deformative strain in the neutral state. b Normal craniocervical junction in flexion. The neuraxis stretches by approximately 10% of its total length with flexion of the craniocervical junction areating a strain z = 0.1. c Pathological craniocervical junction with an abnormal CXA in flexion. Upon full flexion at the craniocervical mice the increase in the tangent are creates a deformative strain approaching z = 0.2 (i.e., 20% stretch). In vivo and in vitro models demonstrate decreased or loss of neurological function with strains of 0.2



Fraser C. Henderson Sr^{1,2} · Fraser C. Henderson Jr³ · William A. Wilson IV² · Alexander S. Mark⁴ · Myles Koby¹

CCA, also known as Clivus Canal Angle and Clivo-Axial Angle As radiologists, we often look for compression of a structure to explain patient symptomatology.

However, there is growing recognition of symptomatic brainstem and/or upper spinal cord deformity with or without compression, particularly within neurosurgical literature.

• Kyphotic CCA is an important metric for predicting the risk of symptomatic brainstem and upper spinal cord deformity and as a useful marker indicating craniocervical instability

• Key Point: Head and neck radiologist need to assess CCA even in the absence of compression.



Chiari Malformations

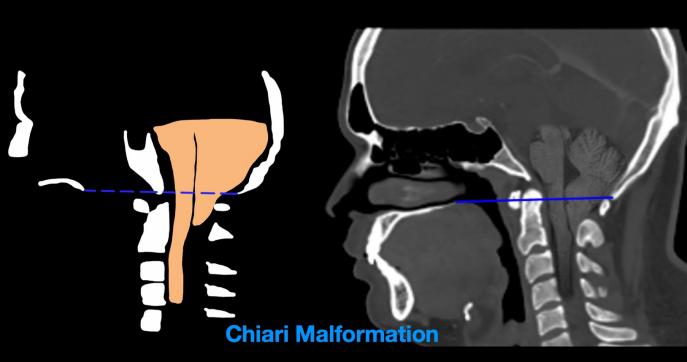
Congenital caudal displacement of the brainstem and cerebellum

Chiari I Malformation

- Chiari 0
 - Syrinx without tonsillar ectopia
- Chiari I
 - Peg-like cerebellar tonsillar ectopia inferiorly through the foramen magr
 - Often associated with a syrinx
- Chiari 1.5
 - Variant of Chiari I, aka Bulbar variant of Chiari I Chiari II

Chiari II

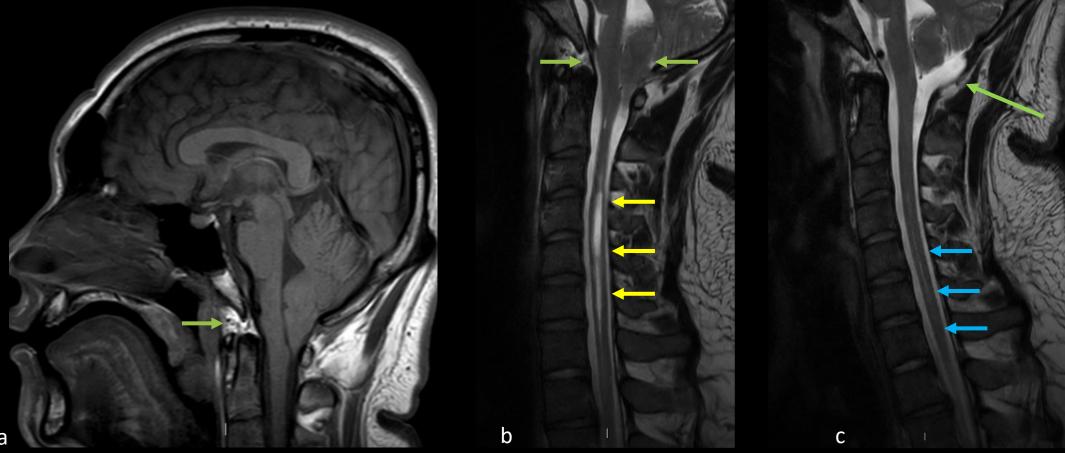
- Lumbosacral myelomeningocele, and inferior ectopia of brainstem, ton and vermis
- Chiari III
 - Chiari II plus low occipital/high cervical encephalocele
- Chiari IV
 - This term is now obsolete.
- Chiari V
 - Absent cerebellum with occipital lobe herniation through foramen magnum



Adapted from Botelho, et al. J Neurol Neuromed 2(3): 15-19



Pre- and Postoperative Chiari I, Normal CCA



а

Chiari I, Normal CCA.

a. Pre-operative Sagittal T1 MRI brain and b. Sagittal T2 MRI c-spine demonstrate the typical pattern of Chiari I cerebellar ectopia (->) with associated compression and marked crowding at foramen magnum with associated cervical cord syrinx (->). There is a normal CCA.

Key Point: Chair I with normal CCA - it is reasonable to assume that the patient's symptoms are related to compressive physiology at the foramen magnum, and that suboccipital decompression should adequately address the patient's symptomatology.

c. Post-operative sagittal T2 MRI c-spine demonstrates an excellent response to suboccipital decompression with resolution of cerebellar ectopia and compression (->) and near complete cervical cord syrinx resolution (->).



Chiari I, Abnormal CCA

Case #1



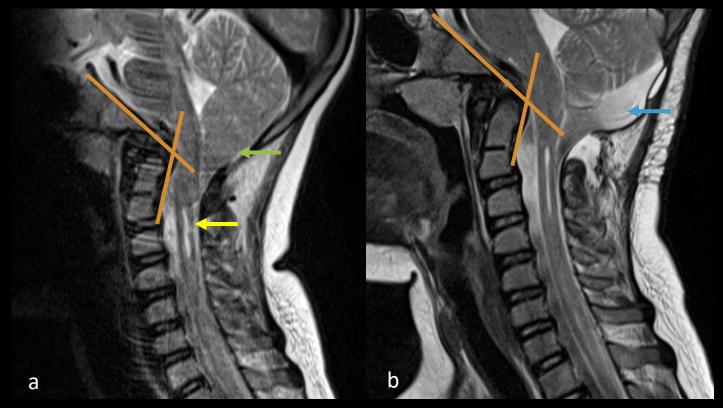
Chiari I, Abnormal CCA.

a. Sagittal T2 MRI C-spine, and b. Sagittal T2-FLAIR and c. Sagittal T1 post-contrast MRI brain demonstrate the typical pattern of Chiari I cerebellar ectopia (->) with associated compression and marked crowding at foramen magnum with associated cervical cord syrinx (->) However, this case is exacerbated by a kyphotic CCA! Actual CCA in neutral position = 132 degrees



Chiari I, Abnormal CCA

Case #2



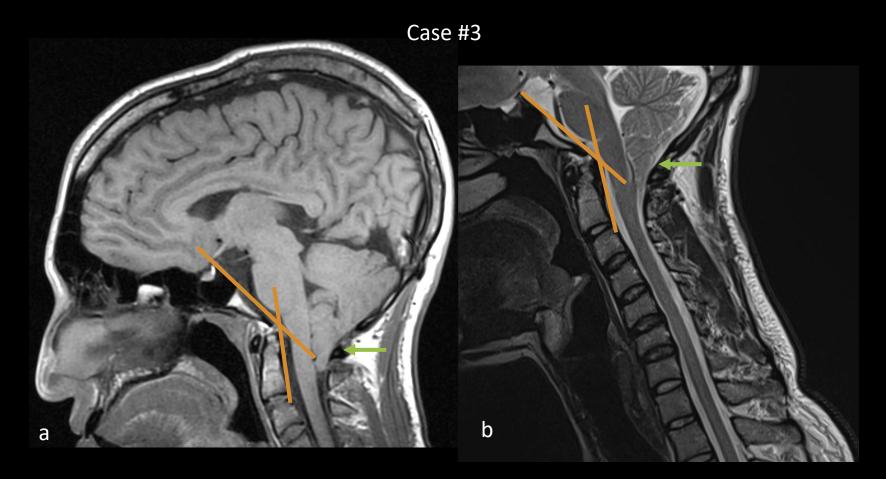
Chiari I, Abnormal CCA.

a. Pre-operative sagittal T2 MRI C-spine demonstrates the typical pattern of Chiari I cerebellar ectopia (->) with associated compression and marked crowding at foramen magnum with associated cervical cord syrinx (->), exacerbated by a kyphotic CCA! = 115 degrees

b. Post-operative sagittal T2 MRI C-spine after sub-occipital decompression surgery (->) shows persistent kyphotic CCA with the patient having persistent CMJ compression symptoms.

Key Point: Chairi I with abnormal CCA - the patient's symptomatology is due to both compression at the CMJ and from deformational stresses from ent of Radiology CCA kyphosis. Decompression alone is unlikely to achieve optimal treatment response! (Henderson et al. Neurosurg Rev. March 2017)

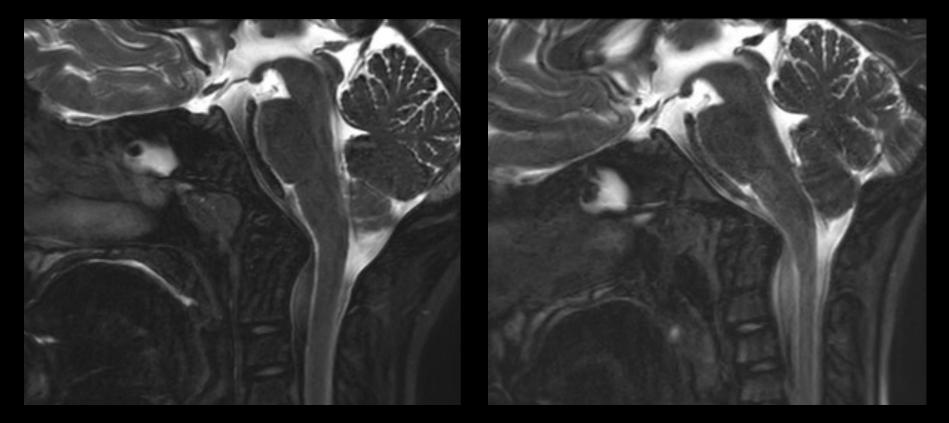
Chiari I, Abnormal CCA



27 year-old male with severe occipital headaches worsened by sneezing, coughing, and laughing.

a. Sagittal T1 MRI Brain demonstrates and b. Sagittal T2 MRI C-spine show Chiari I with cerebellar ectopia (->) with compression and crowding at foramen magnum with associated, exacerbated by a kyphotic CCA of 134 degrees, contributing to the patient's CMJ symptoms which worsen during actions related to head flexion.

Neutral and Flexion MRI

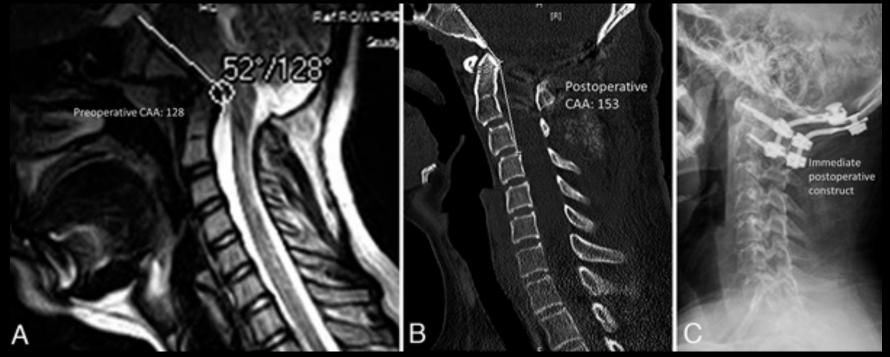


13-y/o with spontaneous micturation every time he flexed his head!

An initial MRI and clinical work-up was negative apart from an abnormal, kyphotic CCA and associated basilar impression. The interpreting radiologist suggested that this may be contributory to the patient's symptoms.

Sagittal STIR MRI in a. neutral position and b. Flexion demonstrates worsening of the CCA kyphosis and increasing basilar impression with flexion relative to the neutral position. Flexion also reproduced the patient's presenting symptoms. The patient underwent corrective surgery.

Chiari I, Kyphotic CCA pre- and post correction



D. Felbaum, et al. J Neurosurg Spine 23:8–15, 2015

A 14-year-old boy, who underwent a suboccipital craniectomy for Chiari Type I malformation 2 years prior continued to suffer from headaches as well as neck and mid-back pain. These occurred on a daily basis and all of his daily activities were limited as a result of his pain. The patient experienced sleep apnea, chronic fatigue, tremor, and insomnia. He exhausted nonsurgical treatments without improvement.

On MRI, he had a CCA of 128 degrees (A). The patient underwent occiput to C-2 fusion. His postoperative CCA measured 153 degrees (B). An immediate postoperative lateral plain radiograph of the cervical spine shows appropriate hardware placement (C).

After OCF surgery, his headaches improved significantly. Additionally, he had decreased tremor, and subjective improvement of his cognition and partment of Radiology attentiveness.

Summary of Key Points

Kyphotic CCA equal to or less than 135 degrees is far more likely to be symptomatic due to ventral cord compression and/or deformation

- Chiari I with normal CCA it is reasonable to assume that the patient symptoms are related to compressive physiology at the foramen magnum, and that suboccipital decompression should adequately address the patients symptomatology
- Chiari I with abnormal CCA the patient's symptomatology is due to both compression at the CMJ and from deformational stresses from CCA kyphosis. Decompression alone is unlikely to achieve optimal treatment response! (Henderson et al. Neurosurg Rev. March 2017)
- Head and neck radiologist need to assess CCA even in the absence of compression.



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